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## SURGERY FOR CONGENITAL HEART DISEASE

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### TWENTY-FIVE-YEAR EXPERIENCE WITH RASTELLI REPAIR FOR TRANSPOSITION OF THE GREAT ARTERIES

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**Objective:** Our purpose was to describe the outcome of the Rastelli repair in D-transposition of the great arteries and to determine the risk factors associated with unfavorable events.

**Methods:** From March 1973 to April 1998, 101 patients with D-transposition of the great arteries and ventricular septal defect underwent a Rastelli type of repair. Median age and weight were 3.1 years (10th to 90th percentiles 0.3-9.9 years) and 12.8 kg (5.9-28.2). Pulmonary stenosis was present in 73 patients and pulmonary atresia in 18; 10 patients had no left ventricular outflow tract obstruction.

**Results:** There were 7 early deaths (7%) and no operative deaths in the last 7 years of the study. Risk factors for early death, by univariable analysis, included straddling tricuspid valve ( $P = .04$ ) and longer aortic crossclamping times ( $P = .04$ ). At a median follow-up of 8.5 years, there were 17 late deaths and 1 patient had undergone heart transplantation. Forty-four patients had

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reoperations for conduit stenosis, 11 for left ventricular outflow tract obstruction, and 28 had interventional catheterization to relieve conduit stenosis. Nine patients had late arrhythmias, and there were 5 sudden deaths. Overall freedom from death or transplantation (Kaplan-Meier) was 82%, 80%, 68%, and 52% at 5, 10, 15, and 20 years, respectively. Freedom from death or reintervention (catheterization or surgical treatment) was 53%, 24%, and 21% at 5, 10, and 15 years of follow-up, respectively.

**Conclusions:** The Rastelli repair can be performed with low early mortality. However, substantial late morbidity and mortality are associated with conduit obstruction, left ventricular outflow tract obstruction, and arrhythmia. (J Thorac Cardiovasc Surg 2000;120:211-23)

Over the past 3 decades the operation described by Gian Carlo Rastelli and his colleagues<sup>1,2</sup> in 1969 has been considered the procedure of choice for the surgical repair of dextro-transposition of the great arteries (D-TGA) associated with ventricular septal defect (VSD) and left ventricular outflow tract obstruction (LVOTO). In the original description, the procedure involved baffling of the VSD to the aorta and connection of the right ventricle to the pulmonary arteries with a homograft conduit. This operation was the first described with the theoretic advantage of incorporating the left ventricle as the systemic ventricle for correction of TGA, and it was widely applied to treat this subset of patients. Its use was also expanded to other anatomic variants of TGA, such as TGA and VSD without pulmonary stenosis as well as double-outlet right ventricle (DORV) until the arterial switch operation was described.<sup>3</sup> After 45 years of surgery for congenital heart disease, several different procedures have been described to treat the subset of patients with complex TGA with or without LVOTO: the Mustard or Senning operation with VSD closure, resection of the LVOTO and/or conduit interposition,<sup>4,5</sup> the arterial switch operation with resection of the LVOTO,<sup>6,7</sup> the *réparation à l'étage ventriculaire*,<sup>8</sup> the Nikaidoh-Bex operation,<sup>9</sup> and a combination of the Ross-Konno switch procedures.<sup>10</sup> Nevertheless, the operation originally described by Rastelli continues to be the most common choice worldwide.

Several reports from different institutions<sup>11-15</sup> have addressed the early and intermediate results of this operation, but little information is available regarding the factors that influence early mortality, late mortality, and freedom from other unfavorable events. In this study we have analyzed our experience with the Rastelli operation specifically for TGA over a 25-year period at Children's Hospital, Boston.

## Patients and methods

**Patient selection.** The pediatric cardiology and cardiac surgery databases and the records of the Department of Cardiac Surgery, Children's Hospital, were extensively searched for patients who underwent any corrective operation for TGA with VSD and LVOTO. A total of 138 patients were identified. Forty-one of this cohort had undergone corrective operations other than the Rastelli procedure, such as Mustard or Senning operations with resection of LVOTO and/or left ventricular-pulmonary arterial conduit, arterial switch operation and resection of LVOTO, Fontan procedure, and a combination of the Ross-Konno switch procedures. Patients who underwent operations other than the Rastelli procedure and patients who underwent Rastelli repair for lesions other than TGA, such as DORV with transposed aorta (n = 6), were excluded from this cohort. A total of 91 patients with TGA, VSD, and LVOTO and 10 patients (8 with previous pulmonary artery banding) with TGA and VSD underwent a Rastelli procedure in the period covered by this study. Thus, 101 patients comprise the study group.

**Definition of D-TGA versus DORV.** To perform as accurate an analysis as possible in terms of morphology, we excluded from this study patients with other diagnoses, such as DORV with D-TGA. We believe that those patients represent another anatomic subset and the surgical technique of VSD baffling is different. The diagnostic landmarks that were used to differentiate D-TGA from DORV with transposed aorta were the presence of a subaortic and subpulmonary conus (thus noncontinuity between the mitral and pulmonary valves), alignment of the pulmonary root relative to the ventricular septum, and coronary pattern.<sup>16</sup>

The study protocol was approved by The Committee on Clinical Investigations of the Children's Hospital.

**Study group.** From March 1973 to April 1998, 101 consecutive patients underwent Rastelli repair for TGA. The segmental anatomy was {S,D,D} in 93 cases, {S,D,L} in 7, and {L,L,L} in 1. Previous studies from our institution have described the results of this operation: first, in 1977, with 7 patients,<sup>17</sup> and then, in 1994,<sup>18</sup> with 11 infants. Those patients

**Table I.** Summary of patients' characteristics and early mortality

Variable	No. of patients	Early deaths	Percent (95% CI)
LVOTO	91	5	5 (2-12)
Pulmonary stenosis	73	5	7 (2-15)
Pulmonary atresia	18	0	0 (0-19)
Multiple VSDs	4	0	0 (0-60)
Inlet extension of VSD	3	0	0 (0-71)
Restrictive VSD	25	1	4 (0-20)
Straddling tricuspid valve	5	2	40 (5-85)
Straddling mitral valve	3	1	33 (1-91)
Cleft mitral valve	6	1	17 (0-64)
Age group			
<1 year	24	3	13 (3-32)
1-11 years	69	4	6 (2-14)
12+ years	8	0	0 (0-37)

CI, Confidence interval.

are included in this series. The study group comprised 39 female and 62 male patients. Patient characteristics are summarized in Table I. Three study periods were defined arbitrarily: 1973-1979 (n = 14), 1980-1989 (n = 46), and 1990-1998 (n = 41). Overall median age and weight at operation were 3.1 years (10th-90th percentiles: 0.3-9.9 years) and 12.8 kg (5.9-28.2 kg).

**LVOTO.** Pulmonary atresia was present in 18 patients and pulmonary stenosis in 73 (29 subvalvular, 34 valvular and subvalvular, 10 valvular). The pulmonary anulus was of normal size in 38 patients (Z-value between -2 and +2) and hypoplastic in 44 (Z-value < -2); pulmonary atresia was found in 19 patients. The pulmonary valve was normal in 29 patients (Z-value between -2 and +2), bicuspid and stenotic in 21 patients, tricuspid and stenotic in 14, doming valve in 19, and atretic in 18. Review of records and angiograms revealed that among 91 patients with LVOTO, 18 did not have anatomic LVOTO as neonates.

**Atrioventricular (AV) valve anomalies.** Five patients had straddling tricuspid valve or abnormal septal attachments, 3 had straddling mitral valve, and 6 had cleft mitral valve.

One patient with TGA and VSD, in whom right ventricular failure and tricuspid regurgitation developed after a Senning operation and VSD closure in infancy, underwent conversion to a Rastelli repair.

**VSD.** Ninety-seven patients had a single VSD and 4 had multiple VSDs (one large conoventricular defect and one or more muscular defects). The VSD was conoventricular in all patients, with an inlet extension in 4 patients. The VSD was found to be nonrestrictive in 76 patients and restrictive in the remaining 25.

**Previous operations.** One or more previous palliative procedures had been performed in 71 patients (Table II).

**Surgical technique.** All operations were performed via a midline sternotomy. Conventional cardiopulmonary bypass (CPB) with bicaval cannulation was used in all patients. The median CPB time was 150 minutes (10th-90th percentiles: 119-225 minutes). In 17 patients for whom a period of circu-

**Table II.** Previous procedures

Type of procedure	n
Blalock-Taussig shunt	49
Blalock-Taussig shunt and Blalock-Hanlon septectomy	4
Balloon atrial septostomy	30
Blalock-Hanlon septectomy	2
Central shunt	3
Pulmonary artery band	2
Pulmonary artery band and Blalock-Hanlon septectomy	2
Pulmonary artery band and coarctation repair	4
Potts shunt	1
Waterston anastomosis	2
Waterston anastomosis and Blalock-Hanlon septectomy	1
Senning repair	1

latory arrest was used, the median time was 53 minutes (12-90 minutes). The median aortic crossclamp time was 87 minutes (59-120 minutes). Fibrillation was used to perform 1 early operation. All operations were performed with the use of hypothermia with a mean rectal temperature of 23.8°C.

After a right ventriculotomy the VSD was identified. When the surgeon considered it necessary, VSD enlargement was performed at the superoanterior margin of the defect. The VSD was baffled to the aorta with pledget-supported interrupted sutures in all but 4 patients. The VSD was enlarged in 47 patients and baffled with a Dacron patch in 89 patients, polytetrafluoroethylene\* in 2, and pericardium in 8.

Right ventricular-pulmonary arterial continuity was achieved with the use of a conduit or by direct anastomosis, in an end-to-side fashion in 9 patients and in an end-to-end fashion in 92. The pulmonary trunk was oversewn in 92 patients and ligated in 9. Direct anastomoses were per-

\*Gore-Tex patch; registered trademark of W. L. Gore & Associates, Inc, Flagstaff, Ariz.

formed without the Lecompte maneuver with the use of the pulmonary artery posteriorly and a polytetrafluoroethylene patch anteriorly. Right ventricular–pulmonary arterial connections were created with the use of 39 aortic homografts, 18 pulmonary homografts, 18 Hancock conduits (Medtronic, Inc, Minneapolis, Minn), 11 Carpentier-Edwards conduits (Baxter Healthcare Corp, Edwards Division, Santa Ana, Calif), 6 Dacron conduits, 4 Tascon conduits (Medtronic Heart Valves, Irvine, Calif), and 5 right ventricular–pulmonary arterial direct anastomoses. The conduit was positioned to the right side of the aorta in 7 patients and to the left side in 94.

One patient received a classic Glenn shunt in conjunction with the Rastelli procedure.

**Early morbidity and mortality.** The postoperative course and autopsy records were evaluated when available. *Early mortality* was defined as death within 30 days after the operation. *Early morbidity* was defined as the presence of more than one of the following: intensive care unit stay of more than 4 days, assisted ventilation for more than 3 days, epinephrine infusion, pleural effusions, renal failure, cardiac arrest, and readmission within 30 days.

**Follow-up.** All 94 hospital survivors could be traced after hospital discharge. A questionnaire was mailed to the patients or parents and the primary physician of patients being followed up at other institutions. Internet databases such as <http://www.info-space.com> or <http://ssdi.advance/ancestry.com> were used to trace addresses for physicians or patients and to determine whether a patient was alive according to the Social Security records to avoid contacting the family of a deceased patient.

**Reoperations.** *Reoperation for right ventricular outflow tract obstruction (RVOTO)* was defined as any procedure that involved relief of RVOTO with or without concomitant procedures such as residual VSD closure, LVOTO reoperation, or mitral valve plasty. LVOTO reoperation and VSD reoperation were determined similarly.

**Data analysis.** Early outcomes for this study were the binary variables early morbidity and early mortality. Relationships between early outcome and categorical perioperative factors were evaluated by the Fisher exact test. When appropriate, subgroups of patients were combined. For continuous perioperative variables, the Wilcoxon rank sum test was used to compare median values for those who were treated successfully and those who were not. To assess the simultaneous effects of perioperative characteristics on early outcome, we considered variables significant at the level in univariable analysis for inclusion in a multivariable logistic regression model. A significance level of .05 was required for retention in the final model.

Late outcomes for the study are time from the Rastelli procedure to death or cardiac transplantation, time to reintervention for RVOTO, and time to reintervention for LVOTO. The effect of early mortality on these late outcomes was eliminated by defining survival time as the period beginning 30 days after the Rastelli procedure. Patients who did not experience a failure were considered to be censored at the time of last follow-up. Survival estimates were obtained by the Kaplan-

Meier method. Distributions of survival times for subgroups of patients defined by the categorical perioperative factors were compared by the log-rank test. The Cox proportional hazards model was used to assess the effects of continuous factors and to perform multivariable analyses.

## Results

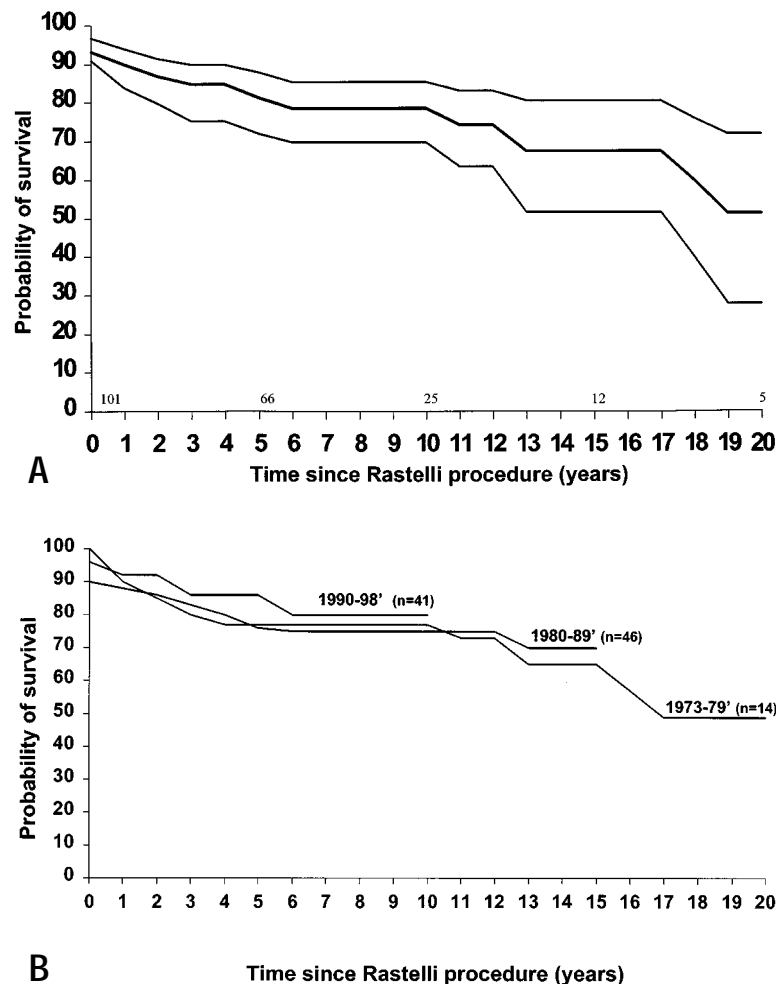
**Overall survival.** Survival at 1 month and at 5, 10, 15, and 20 years was 93%, 82%, 80%, 68%, and 52%, respectively (Fig 1, A).

**Early mortality.** Early mortality was 0% in 1973-1979 (95% confidence interval [CI] = 0%-23%), 11% in 1980-1989 (95% CI = 4%-24%), and 5% in 1990-1998 (95% CI = 0.6%-17%). There were no early deaths in the most recent 7-year period. There were no statistically significant differences among the 3 periods with respect to survival ( $P = .8$ ). The cause of death was found to be related to residual LVOTO in 3 cases, coronary insufficiency in 2, sudden AV block not responsive to pacemaker stimulation in 1, and pulmonary hypertension in 1.

Age group (<1 year, 1-11 years, 12+ years) and weight were not significant risk factors for early death ( $P = .5$  and  $P = .2$ , respectively). In univariable analysis, the only significant morphologic risk factor for early mortality was the presence of a straddling tricuspid valve ( $P = .04$ ), which occurred in 5 patients, 2 of whom died early (Table III). Procedural risk factors included longer aortic crossclamp time ( $P = .04$ ). Patients with Carpentier-Edwards conduits were more likely to die early ( $P = .03$ ). Although early mortality was more prevalent in patients with mitral valve anomalies (22% vs 5%,  $P = .12$ ), this relationship did not achieve statistical significance.

Multivariable analysis was unable to detect any of these factors that resulted in a greater likelihood of early death. An association was noted between early mortality and the postoperative variable complete AV block that did not resolve until death or discharge with pacemaker implantation; among the 6 patients in whom complete AV block was noted, 3 died early ( $P = .004$ ). There was also an association between longer CPB time and early mortality ( $P = .02$ ).

**Late mortality.** At a median follow-up of 8.5 years (range 0.4-22 years), there were 17 late deaths and 1 heart transplantation. The causes of late death or failures are summarized in Table IV. Survival for the 3 periods is summarized in Fig 1, B; there are no significant differences over the 3 study periods ( $P = .9$ ). In univariable analysis, the risk factors for shorter time to late death or failure were straddling tricuspid valve ( $n = 3$  patients,  $P < .001$ ), use of a pericardial baffle ( $P =$



**Fig 1. A,** Overall survival after the Rastelli repair, with 95% confidence limits. **B,** Survival for the 3 study periods.

.02), length of stay more than 14 days ( $P = .03$ ), and use of circulatory arrest ( $P = .06$ ). Use of a pericardial baffle (hazard ratio = 7.5, 95% CI = 1.5-38,  $P = .01$ ) and length of stay more than 14 days (hazard ratio = 3.2, 95% CI = 1.2-8.4,  $P = .02$ ) remain significant in multivariable analysis.

**Hospital morbidity.** Early morbidity had a prevalence of 40% (40/101). The median length of hospital stay, intensive care unit stay, and ventilatory support among hospital survivors were 9 days (6-21 days, 10th and 90th percentiles), 3 days (2-8 days), and 1 day (1-3 days), respectively. There were 10 early reoperations, 4 for bleeding, 3 for pacemaker implantation, 2 for placement of left ventricular apex–descending aorta conduits (both patients died within hours of reoperation), and 1 for creation of a pericardial window in a patient with recalcitrant pericardial effusions. One patient required interventional catheterization to retrieve a retained

catheter fragment from the left atrium. A 3-month-old boy received support with an extracorporeal membrane oxygenator after cardiac arrest in the catheterization laboratory and was successfully weaned immediately after the Rastelli procedure. Sternal closure was delayed in 5 patients, 1 of whom died.

In univariable analysis, the procedural risk factors for early morbidity were any prior surgery (46% vs 23%,  $P = .04$ ), previous coarctation repair (100% vs 37%,  $P = .02$ ,  $n = 4$  patients who had coarctation repair), use of total circulatory arrest (76% vs 32%,  $P = .001$ ), and right-sided conduit (86% vs 36%,  $P = .02$ ). Although patients with mitral valve anomalies were more likely to experience early morbidity (67% vs 37%,  $P = .15$ ), this relationship did not achieve statistical significance. Associations were also found between early morbidity and the postoperative variable AV block ( $P = .04$ ) and longer CPB time ( $P = .02$ ). In a multivariable analysis,

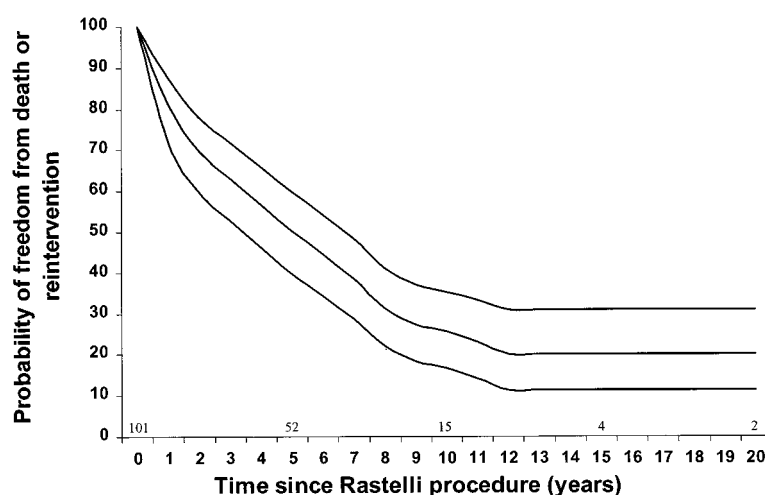


Fig 2. Freedom from reintervention, reoperation, or death.

**Table III.** Perioperative variables associated with early mortality in univariable analysis

Variable	Early mortality		P value
	Yes (n = 7)	No (n = 94)	
Straddling tricuspid valve	2 (29%)	3 (3%)	.04
Carpentier-Edwards conduit	3 (43%)	8 (9%)	.03
Aortic crossclamp time (min)	100 (88-120)	86 (67.5-99.5)	.04

Values are presented as number (percent) for categorical perioperative variables and median (interquartile range) for continuous variables.

patients were more likely to experience early morbidity if they had any use of circulatory arrest (odds ratio = 9.1, 95% CI = 2.4-34,  $P = .001$ ) or any prior operation (odds ratio = 3.9, 95% CI = 1.3-12,  $P = .02$ ).

**Arrhythmia and AV block.** Twenty-four patients (24%) had 33 episodes of postoperative arrhythmia. Junctional ectopic tachycardia was seen in 11 patients, supraventricular tachycardia in 9, complete AV block (AV block that did not resolve until pacemaker implantation or death) in 6, transient AV block (AV block on the day of operation) in 5, and ventricular tachycardia in 2. Of 6 patients with complete AV block, 3 died early. The remaining 3 received permanent pacemakers. There was no relationship between VSD enlargement and either complete or transient AV block.

**Late morbidity.** Freedom from death or reintervention (catheterization or surgical) was 53%, 24%, and 21% at 5, 10, and 15 years of follow-up, respectively (Fig 2). There were 53 first reoperations, 12 second, 5 third, 2 fourth, and 1 fifth. Interventional catheteriza-

**Table IV.** Causes of late death or failure

Causes	n
Sudden death	5
Left ventricular dysfunction	7
Conduit pseudointima rupture	1
Myocarditis	1
Unknown	2
At reoperation	2

tion was performed on 36 occasions in 28 patients. Five reoperations and 1 interventional catheterization were performed at other institutions. Two patients died at reoperation (1 had VSD closure with aortic valve replacement, and 1 had conduit replacement).

**RVOTO.** Sixty-four of 94 survivors had at least mild RVOTO during the follow-up period. Freedom from reintervention for right-sided obstruction was 56%, 25%, and 21% at 5, 10, and 15 years, respectively (Fig 3).

Among 44 patients with reoperations for conduit stenosis, 10 underwent concomitant resection of hypertrophied right ventricular muscle bundles. In 34 patients the conduit was changed for another conduit (11 pulmonary homografts, 9 aortic homografts, 3 Carpentier-Edwards conduits, 2 Hancock conduits, 2 polytetrafluoroethylene conduits, and 7 Dacron conduits), and in 10 an onlay patch or patch augmentation was used.<sup>19</sup> One patient with a calcified aortic homograft had a massive hemorrhage from the aorta at the

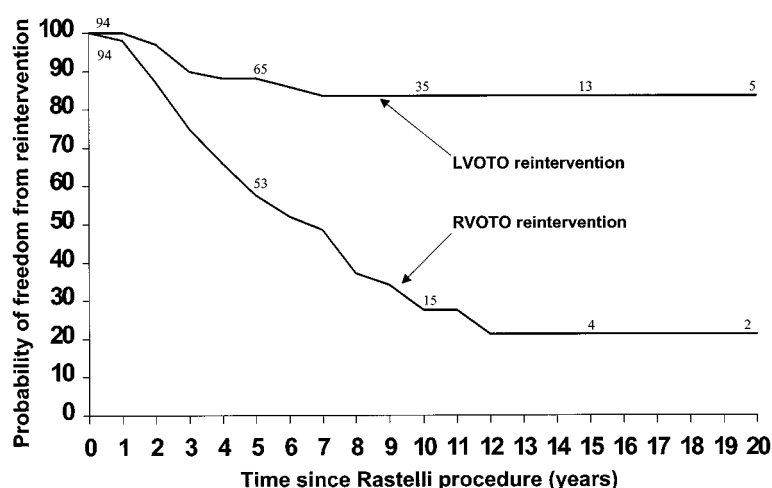


Fig 3. Freedom from RVOTO and LVOTO reintervention.

time of sternotomy and died 5 days later of neurologic complications. Time to RVOTO reintervention was shorter for patients less than 1 year of age at the original operation ( $P < .001$ ) and for those with right-sided conduit placement ( $P = .02$ ). Time to reintervention did not differ by the type of right ventricular–pulmonary arterial connection ( $P = .2$ ), although no patient with a direct anastomosis ( $n = 5$ ) underwent a reintervention.

**RVOTO transcatheter reintervention.** Since 1984, 28 patients have undergone transcatheter reinterventions. Sixteen patients received 1 or more vascular stents and 12 received balloon dilation alone. Conduits treated include 18 aortic homografts, 6 pulmonary homografts, 2 Tascon, 1 Carpentier-Edwards, 1 Dacron, and 1 Hancock. The gradient in obstructed conduits was reduced from  $66.4 \pm 14.7$  mm Hg to  $28.4 \pm 11.8$  mm Hg ( $P = .001$ , paired  $t$  test) and the diameter at the obstruction was increased from  $7.8 \pm 3.6$  mm to  $12.7 \pm 2.9$  mm ( $P = .01$ ). Ten patients underwent a second reintervention and 3 a third. The freedom from surgical reintervention after transcatheter reintervention was 65% and 22% at 2 years and 5 years, respectively.

**LVOTO reintervention.** Eleven patients underwent surgical revision of LVOTO with no early deaths. Nine patients had resection of the conal septum, 1 patient a modified Konno procedure, and 1 had an apical left ventricular–descending aorta conduit (died 3 years later). One patient underwent stent placement in the LVOT. Freedom from an LVOTO reintervention was 88%, 84%, and 84% at 5, 10, and 15 years, respectively (Fig 3). Time to LVOTO reintervention was somewhat shorter for infants less than 1 year of age at the Rastelli repair ( $P = .08$ ).

**Other procedures.** Residual VSD closure was performed as a solitary procedure in 3 patients. Closure of residual VSD was performed concomitant with the conduit reoperation in 11 patients. One patient had device closure of a residual VSD. Two patients underwent mitral valve cleft closure, 1 a mitral valve replacement, and 1 an aortic valve replacement with residual VSD closure.

**Late arrhythmia.** All patients had right bundle branch block at late electrocardiographic examination. At follow-up, AV block was present in 5 patients (3 after surgery and 2 acquired late), supraventricular tachycardia in 4 patients, and ventricular tachycardia in 3. There were 5 sudden deaths. Two patients underwent pacemaker implantation and 2 patients received an automatic implantable cardiac defibrillator.

**Pregnancies and deliveries.** Two patients had 3 pregnancies and deliveries without complications.

## Discussion

**Early morbidity and mortality.** The surgical treatment for TGA has evolved dramatically over the past 40 years, and currently the procedures to treat this anomaly have an operative mortality of less than 5%. Over the 25 years encompassed by this study, the policy of primary repair that has characterized our institution was also applied for this anomaly. However, in this series mortality was not influenced by earlier date of operation. In fact, during the first 8 years of this experience there were no early deaths.<sup>17</sup>

In this series the only morphologic risk factor for early mortality was the presence of tricuspid valve straddling or abnormal attachments (3 deaths in 5

patients), where VSD baffling becomes more difficult. Under such circumstances, it is necessary to divide the chordae and attach them to the VSD baffle. Niinami and colleagues<sup>20</sup> have described a technique to avoid the complications of tricuspid malinsertion in this subset. Another point for consideration is the common association of right ventricular hypoplasia and straddling tricuspid valve. In this setting, it may be appropriate to include a bidirectional Glenn shunt in the correction with the theoretic advantage of decreasing the right ventricular volume load.<sup>21</sup>

Additional risk factors for death were the use of a Carpentier-Edwards conduit and a right-sided conduit. This association may be related to the potential problem of coronary compression by the prosthetic valve ring.<sup>22</sup> In this series, 7 patients had an L-transposed aorta, necessitating a right-sided conduit. The relation of the conduit with the sternum is crucial in a Rastelli repair. Right-sided conduits may be more affected by sternal compression since the position of the right ventricular infundibulum is more anterior.<sup>23</sup>

**RVOTO.** The late results of heterograft valve conduits<sup>24</sup> are similar to those of homografts.<sup>25</sup> However, the use of heterografts such as the Carpentier-Edwards conduit was associated with early death in this series. This poor result may be due to disadvantages with respect to bleeding, coronary compression, and difficulty suturing to the thin and friable pulmonary arteries and ventricle. Also, replacement of an obstructed conduit in a Rastelli repair can be a difficult procedure, particularly if it is adherent to the sternum. In such cases, groin cannulation before resternotomy can be a lifesaving procedure.

The late results of the direct anastomosis of the right ventricle to the pulmonary artery are encouraging since no intervention was required in the late follow-up. However, its use was abandoned in our unit after a patient with DORV experienced intermittent left coronary artery compression from the tethered right pulmonary artery.

Another approach for RVOT reconstruction is to perform a Lecompte maneuver.<sup>15</sup> However, in TGA, VSD, and LVOTO, the ascending aorta is large and, unlike repair of simple TGA, is not reimplanted posteriorly. The presence of branch pulmonary artery stenosis in combination with free pulmonary regurgitation may lead to severe postoperative right ventricular failure. Therefore, the use of the Lecompte maneuver or the French modification of the Rastelli procedure (*réparation à l'étage ventriculaire*) should be reserved for patients with nearly normal sized ascending aortas in D-TGA or for patients with side-by-side great vessels.

In the past few years, interventional catheterization procedures have been used successfully to prolong the life span of the right ventricular-pulmonary arterial conduit at our institution. In general, implantation of a stent in the right ventricular-pulmonary arterial conduit is considered when right ventricular pressure exceeds 75% of the systemic pressure or in the presence of right ventricular dysfunction or dilatation and lower right ventricular pressures. Most commonly, obstruction occurs at the level of the homograft valve, which is typically stenotic, calcified, and regurgitant. In such cases, stent placement usually results in free pulmonary regurgitation. In the presence of distal branch pulmonary artery stenoses or hypertension, stent implantation in such positions is avoided, and surgical conduit replacement is preferred. When the obstruction is located in the middle of the homograft or in the distal anastomosis, stent implantation is attempted distal to the homograft valve.<sup>26</sup> In addition, stent implantation is not considered if it would at most increase the diameter 1 or 2 mm, unlikely to change the pressure gradient appreciably.

**LVOTO.** The technique of VSD baffling for the Rastelli procedure has evolved in our experience, and in the past 10 years enlargement of the VSD is done almost routinely. Rychik, Jacobs, and Norwood<sup>27</sup> have postulated that in TGA, as in double-inlet left ventricle, the VSD or bulboventricular foramen becomes restrictive after the biventricular repair or the Fontan operation because the left ventricular volume is often increased before the correction. However, resection of the anterosuperior margin of the defect carries a potential increased risk for AV block and scar tissue as a substrate for arrhythmia. The presence of hypertension in the right ventricle would be another factor for late LVOTO, since right ventricular hypertension will produce septal hypertrophy and leftward septal displacement. The tunnel shape of the LVOT after a Rastelli repair may be affected by right ventricular hypertension.

**Late failure, mortality, and arrhythmia.** The survival at 5, 10, and 15 years seems disappointing in comparison with late results reported after atrial or arterial repairs of TGA or even the Fontan procedure. The analysis of causes of late death revealed that left ventricular failure and sudden death play an important role.

In a study by Graham and coworkers,<sup>28</sup> abnormal left ventricular wall stress was demonstrated in patients who underwent a Rastelli procedure. In that study the mean end-systolic stress and the left ventricular mass were elevated and contractile function was abnormal in



8 of 11 patients. This information indicates that left ventricular function is frequently abnormal and residual left ventricular dilation and wall hypertrophy remain despite “successful” application of the Rastelli operation. However, the age of those patients at operation was considerably higher than the mean age during the period 1990-1998 in our experience. Hypoxia, obstruction, and volume overloading over a prolonged period may explain the contractile function reported. Other factors that may affect contractile function are an abnormal interventricular septum with a large prosthetic component (the VSD baffle) and the presence of right ventricular hypertension impairing left ventricular filling and leftward septal displacement.<sup>29</sup>

The relatively high incidence of late arrhythmia is likely caused by right bundle branch block that may progress to bifascicular and complete AV block, scar tissue, and right ventricular hypertension due to an obstructed conduit. Sudden death seen in our study may be attributed to 2 causes: sudden ventricular fibrillation or sudden AV block. Holter monitoring with pacemaker and/or automatic cardiac defibrillator implantation, when indicated, may decrease the incidence of late sudden death.<sup>30</sup>

**Indications for the Rastelli procedure in the current era.** The optimal age for the performance of a Rastelli repair remains controversial because of the balance between palliation and correction. Palliation leads to the performance of Rastelli repair at an older age with larger conduits, therefore reducing the need for reoperations. However, palliation also leads to hemodynamic deterioration, overloading, and some degree of residual cyanosis. Since 1987 the philosophy of early repair was adopted for this subset at our institution to avoid the deleterious effects of cyanosis and ventricular overloading.

In 1969, the Rastelli procedure<sup>1,2</sup> became the first successful operation for TGA that incorporated a systemic left ventricle and therefore was called *anatomic* correction. However, the Rastelli procedure, an epic landmark of pioneering in cardiac surgery, is far from being an anatomic correction since the right ventricle is connected to the pulmonary artery with a prosthetic conduit that in most cases will become obstructed, and the LVOT is created by baffling the VSD with another prosthesis. The real anatomic correction for TGA and VSD was first described in 1975,<sup>3</sup> but this procedure had its learning curve and it was not widely used in the first years. The current excellent early and late results of the arterial switch operation have relegated the Rastelli procedure to an alternative when LVOTO is not amenable to relief at the time of arterial switch.

LVOTO in TGA is commonly associated with posterior deviation of the conal septum, defined as the muscular structure between the arterial roots that divides the conus to subaortic and subpulmonary portions.<sup>16</sup> The posterior malalignment of the conal septum creates a narrowing to the subpulmonary area that becomes more obstructive with time.<sup>31</sup> Interestingly, 18 patients of the group with TGA, VSD, and LVOTO in this study had no anatomic LVOTO when they were newborn infants. Furthermore, 29 had a normal pulmonary valve. In our study population, the severity of LVOTO increased with time. We speculate that the performance of an early arterial switch operation might avoid the development of LVOTO in many patients. The presence of a systemic left ventricle avoids the leftward septal displacement and therefore LVOTO; another postulate is that the VSD patch might produce a rightward-anterior traction of the conal septum. In the arterial switch study of the Congenital Heart Surgeons Society,<sup>32</sup> subaortic stenosis developed in only 2 of 514 patients. After 2 years, left-sided obstruction occurred at a rate of about 0.1% per year. Furthermore, the prevalence of late LVOTO in patients with atrial corrections was around 5%<sup>33</sup> and the prevalence of development of LVOTO was around 9% in a period from birth until the performance of the physiologic (nonanatomic) correction.<sup>34</sup>

The presence of anatomic features that may produce LVOTO by a different mechanism, such as small left ventricular-aortic junction and abnormal mitral valve attachments, may not be influenced by an early arterial switch operation. A cleft mitral valve, abnormal valve attachment to conal septum or papillary muscle, and accessory mitral valve tissue are anatomic features that will not be affected by a dynamic element. If the obstruction in those cases is above the VSD, a Rastelli repair is advisable. The high prevalence of complications in the late follow-up after Rastelli repair and the excellent results of the arterial switch operation suggest that an arterial switch is the preferable operation for a newborn with TGA, a conoventricular septal defect, and dynamic or mild anatomic LVOTO by a membrane amenable to resection.

Furthermore, when a Rastelli procedure is likely to be complicated by increased early and late morbidity and mortality due to morphologic risk factors, alternative procedures such as one and a half ventricle repair, Ross-Konno switch, or even a Fontan procedure should be considered. As reported by Delius and colleagues,<sup>35</sup> a high-risk biventricular repair is not always preferable to a single ventricle repair.

## Study limitation

This study encompasses a period of 25 years during which constant changes in preoperative, operative, and postoperative management have almost surely affected outcomes in a way that is not captured by our analyses. Within this limitation, however, we have identified a base incidence of death and complications that appears to continue immutably throughout the follow-up of patients who have undergone the Rastelli repair.

## Summary of analysis

1. The Rastelli repair has been performed with low early mortality throughout the past 3 decades.
2. The presence of straddling tricuspid valve appears to be a risk factor for early and late mortality although the small number of patients with this anatomic defect makes definitive conclusions tenuous.
3. Complete AV block appears to be associated with early mortality.
4. Despite these early excellent results, late follow-up indicates a high prevalence of conduit obstruction, LVOTO, arrhythmia, and death.
5. The prevalence of early and late arrhythmia is high after Rastelli repair.
6. VSD enlargement by resection of the anterosuperior margin of the defect was not associated with either complete or transient AV block and therefore constitutes an appropriate tool to deal with LVOTO at VSD baffling. Awareness of LVOTO has led us to frequently include VSD enlargement concomitant with the Rastelli procedure.
7. The late results were not different between the various conduits. However, the populations were different for the heterografts and homografts. The only technique of RVOTO reconstruction with good late results was direct anastomosis.

During the first 2 decades of this study, surgical care of these patients was provided by Aldo R. Castañeda, MD, William I. Norwood, MD, and Frank L. Hanley, MD.

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## Appendix: Perioperative variables\*

### Patient-related variables

*Age at repair*  
*Weight at repair*  
 Body surface area at repair  
*Sex*  
 Decade of procedure  
 Diagnosis: TGA+VSD, TGA+VSD+PA,  
 TGA+VSD+PS  
 VSD type  
 Restrictive VSD  
 VSD enlargement  
 Multiple VSDs  
 Presence of anatomic LVOTO as newborn  
 Size of pulmonary anulus  
 Type of pulmonary valve  
 Discontinuity of pulmonary arteries  
 Straddling tricuspid valve  
 Mitral valve anomalies  
 LSVC to coronary sinus  
 Prior surgery: Blalock-Taussig shunt, pulmonary artery  
 band, coarctation repair, Blalock-Hanlon septectomy

### Procedure-related variables

Conduit type: aortic homograft, pulmonary homograft,  
 Carpentier-Edwards conduit, Tascon conduit, Dacron con-  
 duit, direct anastomosis, Hancock conduit  
 Conduit oversizing  
 Use of circulatory arrest  
 Aortic crossclamp time  
 Right-sided or left-sided conduit  
 VSD baffle

### Postoperative variables (for analysis of late outcomes only)

Days receiving ventilatory assistance  
 Duration of ICU stay  
 Duration of hospital stay  
 Complete or transient AV block

## Discussion

**Dr Gordon K. Danielson** (*Rochester, Minn*). [Slide] This photograph was taken of Gian Carlo Rastelli in the year he proposed his procedure, and it was the same year the first clinical case was performed at the Mayo Clinic by Dr Robert Wallace on July 26, 1968. Tragically, Dr Rastelli died of Hodgkin disease less than 2 years after this photograph was taken.

The patient was a 14-year-old boy who had undergone 2 previous Blalock-Taussig shunts. A radiated homograft was used as a conduit, and this was replaced 8 years later with a Hancock conduit. Now, 22 years after that fourth operation,

\*Abbreviations: TGA, transposition of the great arteries; VSD, ventricular septal defect; PA, pulmonary atresia; PS, pulmonary stenosis; LVOTO, left ventricular outflow tract obstruction; LSVC, left superior vena cava; ICU, intensive care unit; AV, atrioventricular.

and nearly 30 years after the Rastelli repair, his health is excellent, he is employed full time, he is receiving no medications, and he has married and has 2 normal children. Clearly, despite some of the complications you have reported, some patients do extremely well after the Rastelli repair.

The authors have shown that straddling tricuspid valve is a risk factor for both early and late mortality. We have encountered another, perhaps more common, anomaly of the tricuspid valve that also presents difficulties in construction of the baffle from the right ventricle to the aorta, this being the tricuspid valve orifice.

The papillary muscle attachments in these anomalies are high, ranging from 9 o'clock to 3 o'clock, and interfere with the direction of the baffle. This is opposed to the usual insertion of the papillary muscles from about 6 o'clock to 8 o'clock near the inferior rim of the defect.

We had previously learned that division of papillary muscles with subsequent reattachment to a septal defect patch was not satisfactory as a solution for us, because later tricuspid insufficiency occurred. In your manuscript you did mention that some papillary muscles had been divided and reattached. Thus, my first question is this: Have you encountered this type of papillary muscle abnormality? If so, how do you place your baffle?

Second, have you seen late tricuspid insufficiency from that division and reattachment of the papillary muscles?

In a review of our late experience with extracardiac conduits, we found that freedom from reoperation for conduit failure was least for the diagnosis of TGA repaired by the Rastelli procedure. Nevertheless, 40% of the patients at 10 years were still free of reoperation for conduit obstruction. This compares with the 10-year freedom from reintervention of only 28% in your series. We believe this could be attributed primarily to the younger mean age of the patients at the time of operation in your series, because you found young age to be a significant risk factor for conduit reintervention. This raises the difficult issue of timing in the placement of an extracardiac conduit.

There also was a trend toward an increased risk in permanent, complete heart block in the younger patients in your study. It is widely known that your group has pioneered in showing that the best treatment for congenital heart defects in most patients is repair in early infancy. What do you now believe is the ideal age for a Rastelli repair of TGA, VSD, and significant subpulmonary stenosis?

**Dr Kreutzer:** First, I would like to emphasize the debt that we all owe to Dr Gian Carlo Rastelli for his great contributions, not only in the field of TGA, but also with truncus arteriosus, tetralogy of Fallot, pulmonary atresia, and the great contributions in the field of common AV canal.

Regarding Dr Danielson's questions, I would like to point out that 5 of our patients had straddling tricuspid valve, and our experience with straddling tricuspid valve in Rastelli operations is very disappointing. The patients who did not die early died late. Straddling mitral valve was a definite risk factor for early and late mortality.

In 2 patients the baffle was constructed below the chordae, so there was no detachment, but in 3 there was detachment. Two of those patients had a mild to moderate degree of tricuspid regurgitation in the late follow-up period.

The straddling tricuspid valve is usually associated with a hypoplastic right ventricular infundibulum. This is a particular disadvantage in a Rastelli patient with a large ventriculotomy, because the infundibulum has become reduced by the fact that the baffle is placed anterior and is actually occupying part of the right ventricular infundibulum. That is another factor in the poor results obtained in patients who have a straddling tricuspid valve.

The optimal age to perform a Rastelli repair remains a subject of controversy. There are both advantages and disadvantages to an early repair in TGA. The advantages are that hypoxia is abolished and overloading of the systemic left ventricle is relieved. Among the disadvantages, the rate of reoperation for a right-sided obstruction is increased. I believe the best timing for repair has not yet been established. There was no statistically significant difference in age at repair and the presence of a left-sided obstruction, although the prevalence of complete AV block after surgery was more common in the last part of the study. I would like to note that even though AV block was more common, the difference did not reach statistical significance.

**Dr Dominique R. Metras (Marseille, France).** These results illustrate very well that even in the hands of the most prestigious group, with excellent early results, the late morbidity due to reoperation is very important in TGA with VSD and pulmonary stenosis. This is particularly true for RVOT reconstruction with a conduit.

Two years ago we reported to this Association another approach to repair the RVOT. It involves constructing a conduit between the right ventricle and the pulmonary arteries with a tubular segment of ascending aortic autograft. End-to-end anastomosis is easily performed in the long ascending aorta, and a tube of living and autologous tissue extends the pulmonary artery to easily reach the right ventriculotomy. It allows growth, and we have seen in 2 babies, operated on at the age of 3 months, who are now 4 years old and have tripled their weight, that they have wide open RVOTs with normal right ventricular pressure. It also allows, we think, primary correction in infancy without a pulmonary shunt and may substantially decrease the reoperation rate for pulmonary stenosis. This autologous tissue connection also allows the orthotopic posterior position of the pulmonary bifurcation as opposed to the *réparation à l'étage ventriculaire*—Lecompte approach, transferring the pulmonary bifurcation anterior to the aorta with potential dangers of compression and traction.

This is also true for the LVOT. We have extensively used the Lecompte approach of extensive resection of the infundibular septum leading to a more direct connection between the VSD and the aorta.

I have a question for Dr Kreutzer concerning the 11 cases of LVOTO reoperation. Were these cases in which you

enlarged the VSD, and do you care in your procedure to resect the infundibular septum between the pulmonary outflow and aortic anulus?

**Dr Kreutzer.** First, let me assure you that I am aware of your technique of reconstruction. I enjoyed your presentation 2 years ago in Washington, DC, at this meeting. I think the technique is very attractive. Dr Jonas has both experimental and clinical experience with that technique. However, the main problem is that there is pulmonary incompetence in the early postoperative period. We all know that early morbidity is influenced by the presence of free pulmonary regurgitation in the early postoperative period.

In regard to the *réparation à l'étage ventriculaire*, I absolutely agree with you. In TGA, VSD, and LVOTO, the size of the ascending aorta sometimes precludes the performance of a *réparation à l'étage ventriculaire* and of a Lecompte maneuver. In cases in which the Lecompte maneuver is performed, the branch pulmonary arteries will be compressed in the posterior aspect of the pulmonary artery by this huge ascending aorta, and the result will be the lethal combination of full branch pulmonary stenosis and free pulmonary regurgitation.

With respect to the third question, the VSD was enlarged mainly in the anterior margin of the defect. Only 2 patients had resection of the infundibular septum.

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